The diagnosis, morphological particularities, and surgical technique in a case of intravascular leiomyoma extended to the right heart chambers

Zoltan Galajda, MD, PhD,[a] **Constantin Copotoiu, MD, PhD,**[b] **Horatiu Suciu, MD, PhD,**[c] **Diana Tint, MD, PhD,**[d] **Tibor Glasz, MD, PhD,**[e] and **Radu Deac, MD, PhD,**[c] *Debrecen, Hungary; Targu Mures, Romania; Brasov, Romania; and Budapest, Hungary*

Intravenous leiomyoma is a benign smooth muscle cell tumor of uterine origin that may grow into the pelvic veins and the inferior vena cava. It usually affects premenopausal women and the majority (90%) are parous. Because cardiac involvement is present in up to 10% of cases, it may be misdiagnosed as a primary cardiac tumor or a venous thrombus-in-transit. We describe a case of intravascular leiomyomatosis with cardiac extension and the morphological particularities of the removed tumor. (J Vasc Surg 2010;51:1000-2.)

CLINICAL SUMMARY

In December 2007, a 40-year-old woman presented at our institution with diagnosis of right atrial myxoma after having experienced mild dyspnea on exertion, palpitations, and syncopes, without signs of pedal edema. The patient had undergone hysterectomy five years earlier for uterine leiomyomatosis. Two years later, salpingooophorectomy was performed.

Because of recurrent syncopes, the patient received urgent open heart surgery for right atrial myxoma. The surgical procedure revealed a tumor arising from the inferior vena cava and filling the right ventricle, without associated thrombus. Resection of the tumor was performed just above the tape of the inferior vena cava, on cardiopulmonary bypass with cardiac arrest.

After the operation, computed tomography and histology confirmed the diagnosis of an intravascular leiomyoma. It showed a filling defect in the left common iliac vein and inferior vena cava. It also revealed a fibrotic node in the inferior lobe of the left lung (perhaps embolization). The patient was discharged from the hospital in January 2008, and she requested to be transferred for investigation in a medical center of another European country. There, in February 2008, computer tomography of the thorax and abdomen revealed an uncertain image of another tumoral mass attached to the ileocecal region. The discovery was followed by a series of investigations concerning this new pathology, without signs of malignancy on positron emission tomography (PET) images. Despite the diagnosis, the patient decided to wait for the surgery. Resection of the pelvic tumoral mass was performed a few months later in June 2008 in the department of abdominal surgery of the above-mentioned medical center. Histology showed leiomyoma and inflamed lymph nodes. The patient was relatively well without major symptoms. Unfortunately, when she returned home, it was discovered that during all

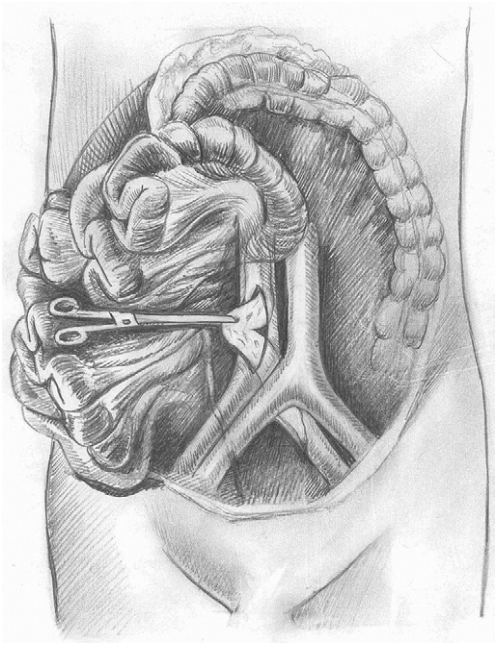

Fig 1. Inferior vena cava is opened and the tumor removed under cardiopulmonary bypass (CPB) assistance.

From the Department of Cardiac and Vascular Surgery, Medical and Health Science Center, University of Debrecen,[a] the First Surgical Clinic, Emergency Clinical Hospital of Targu Mures,[b] the Institute of Cardiovascular Diseases and Transplantation,[c] the Cardiology Department, Faculty of Medicine, Transilvania University,[d] and the Second Department of Pathology, Faculty of Medicine, Semmelweis University.[e]

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Reprint requests: Zoltan Galajda, MD, PhD, Medical and Health Science Center, Departments of Cardiac and Vascular Surgery, Moricz Zsigmond St. 22, 4032 Debrecen, Hungary (e-mail: galajda62@gmail.com).

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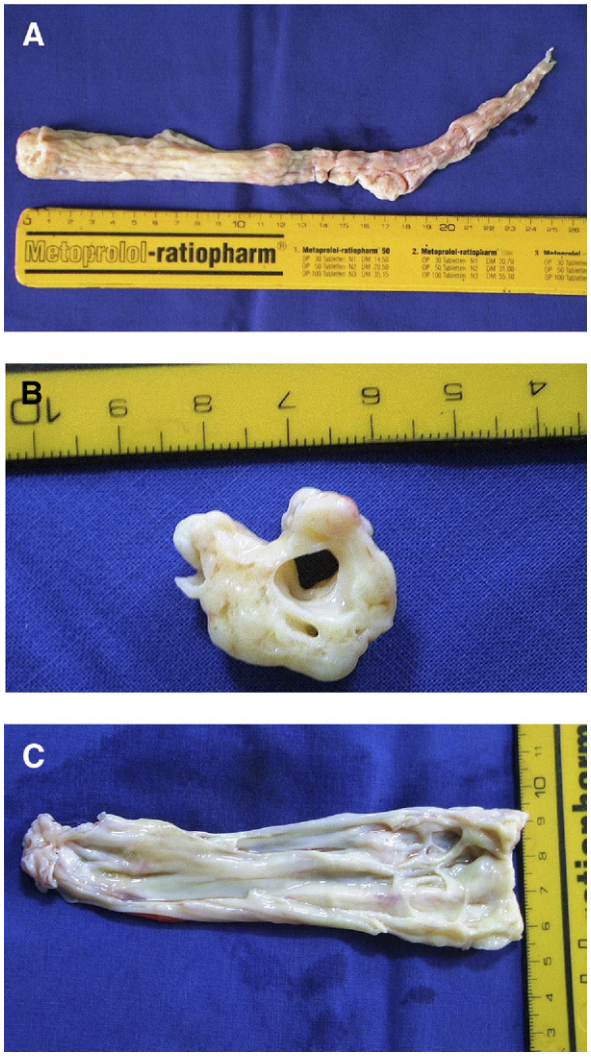

Fig 2. Specimen morphology. *A,* The tumor is 29 cm length arising from left iliac vein to the right atrium. When sectioned transversally *(B)* and longitudinally *(C)*, the tumor showed a lumen inside.

these procedures, the intravascular tumor grew again into the right atrium. We recommended operation in February 2009, and in March 2009, the patient was admitted for surgery in our department. A single-stage procedure was performed via midline sternotomy and laparotomy. We did choose to operate using cardiopulmonary bypass support, because the larger side of the tumor was situated in the right atrium. The two common iliac veins were exposed, as well as the inferior vena cava on a 10 cm length. In parallel, cannulation of ascending aorta, superior vena cava, and left femoral vein were performed. Under normothermic cardiopulmonary bypass (CPB), an oblique atriotomy was performed on the beating heart. A large tumor could be seen arising from the inferior vena cava, which was severely dilated (3 cm) and nearly filled with the tumor. The cava was opened 2 cm cranial from the iliac veins confluence and

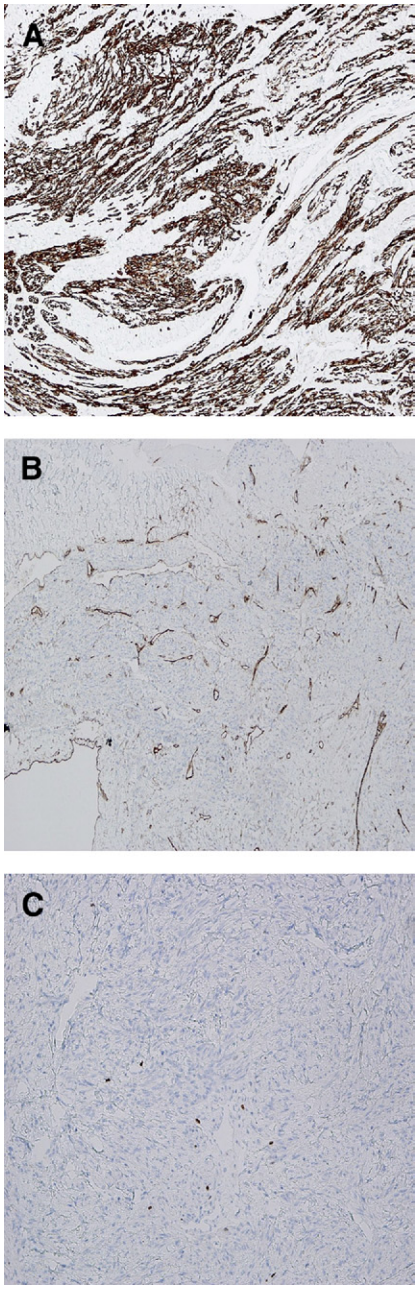

Fig 3. Immunohistochemistry aspects. *A,* Markers for smooth muscle demonstrate myogenic structure of the lesion (SMA, H-caldesmon, 100×). *B,* Immunohistochemstry against endothelium reveals a rich vascular content. (CD34, 100×). *C,* The proliferation marker denotes only scattered positivity (Ki67, 200×).

the tumor was divided on site (Fig 1). Since the upper part of the tumor was not attached, it could be immediately withdrawn through the open inferior vena cava. Under gently blind traction, the inferior part was mobilized from the wall of the left common iliac vein with the help of a dissector instrument. The tumor proved to be attached

only by a thin fibrotic peduncle to an unseen site in the region of internal iliac vein attachment. The total length of the tumor (from the iliac vein to right atrium) was 29 cm, with the presence of an internal, functional lumen, opened in both directions: left common iliac vein and right atrium (Fig 2).

Histological evaluation revealed similar views in both examined tissue samples: in a fibrotic, hyalinized ground tissue, smooth-muscle bundles and scattered single smooth-muscle cells can be seen. Within the larger sample, dilated vascular structures with endothelial lining and well-developed muscular wall layers are found among them. Immunohistochemically with myogenic markers (SMA, h-Caldesmon), the smooth-muscle bundles and the vascular wall structures of both samples became evident (Fig 3, *A*). With the CD-34 endothelial marker, an abundant small-vessel network is shown beside the larger vascular lumens (Fig 3, *B*). Likewise, the proliferation marker Ki-67 decorates less than 1% of all cellular nuclei (Fig 3, *C*).

Based on the findings above, the patient was diagnosed with a benign mesenchymal lesion, the so-called intravascular leiomyomatosis. No change of pulmonary lesion was observed, and, for this reason, the biopsy was not performed.

The patient was discharged one week later without eventful follow-up, and six months after the operation is well, without any complication.

DISCUSSION

Intracardiac leiomyomatosis, a rare condition, generally occurs in women,[1,2] most patients being women of middle age (median age, 44 years).[3] Patients often have a history of hysterectomy. Rarer manifestations that have been reported include a high output state, secondary thrombosis with the Budd-Chiari syndrome, massive ascites, sudden death, and systemic embolism. Metastasis to the lungs and lymph nodes has been reported, and pulmonary nodules have been described. Various lengths of the leiomyoma were reported (15-29 cm in length, 3-3.5 cm in diameter). Recurrence can occur 18 years after surgery.

There are two main theories about the growth mechanism of this tumor: it arises from vascular walls within the myometrium or it is the result of an unusually extensive invasion of the myometrium itself by a leiomyoma.[4]

Immunohistochemical data from literature do not support the hypothesis of a vessel wall origin for intravascular leiomyomatosis.[5] Nevertheless, in our case, the tumor morphology and histology, with existence of an inner circulating lumen, suggest the vascular wall origin. We suppose that this tumor developed as an extension of a vascular wall tumor through the left internal iliac vein. It is difficult to determine histologically the origin of the tumor because the vascular and pelvic tumoral masses were not resected in one-stage procedure. The left common iliac vein was involved from the beginning; it was shown in the first computer tomography images.

In fact, this double circulation (like in aortic dissection) can explain why a tumor of 29 cm in length produced no major embolic events (pulmonary emboly in our case was revealed after the open heart surgery when the atrial tumor was divided and traumatized during cannulation).

Many techniques have been employed in the treatment of tumors of this type.[6] In case of leiomyoma (which is a firm structure resistant to traction), if extended to the inferior vena cava only, a simple laparotomy will then allow the tumor to be removed by downward traction exercised from the iliac vein or the vena cava. In case of leiomyoma extended to the right heart chambers with an estimated larger distal end of the tumor, standby CPB or an abdominal and thoracic approach (sternotomy or right thoracotomy) is recommended, using CPB under normothermy and beating heart.

In our case, the patient was operated on 14 months after the first presentation (multiple concomitant tumors, transfer in other department), but this type of delay in treatment should be avoided.

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AUTHOR CONTRIBUTIONS

Conception and design: ZG, CC, RD, DT
Analysis and interpretation: DT
Data collection: HS, DT
Writing the article: ZG
Critical revision of the article: TG
Final approval of the article: ZG
Statistical analysis: N/A
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Overall responsibility: ZG

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